

Search Articles: 

search tips

HEALTH ARTICLES					MEDICAL COLLEGE PATIENT CARE		
Home	MCW Features	Consumer Information	MCW Columnists	Browse By Topic	Find a Doctor	Clinics & Centers	Request an Appointment

Please Take the  
[HealthLink Survey](#)

## Sandhoff Disease

[Email this article](#)

Sandhoff disease is a rare, genetic, lipid storage disorder resulting in the progressive deterioration of the central nervous system. It is caused by a deficiency of the enzyme hexosaminidase which results in the accumulation of certain fats (lipids) in the brain and other organs of the body. Although Sandhoff disease is a severe form of Tay-Sachs disease--which is prevalent only in people of European Jewish descent--it is not limited to any ethnic group. Onset of the disorder usually occurs at 6 months of age. Symptoms may include motor weakness, startle reaction to sound, early blindness, progressive mental and motor deterioration, frequent respiratory infections, macrocephaly (an abnormally enlarged head), doll-like facial appearance, cherry-red spots, seizures, and myoclonus (shock-like contractions of a muscle).

**Find related articles:**

**By topic:**

[Neurology](#)

**By keywords:**

Receive HealthLink via  
email!

[Subscribe now >>](#)

There is no specific treatment for Sandhoff disease. Treatment is symptomatic and supportive.

The prognosis for individuals with Sandhoff disease is poor. Death usually occurs by age 3 and is generally caused by respiratory infections.

Information provided by the  
**National Institute of Neurological Disorders and Stroke**  
**National Institutes of Health**

*Article Created: 1999-03-20*

*Article Updated: 0000-00-00*

**BEST MEDICAL INFORMATION**  
**BEST MEDICAL INFORMATION**

[Home](#) | [About HealthLink](#) | [Medical College of Wisconsin](#) | [Clinical Trials](#)  
[Contact Information](#) | [Site Map](#) | [Disclaimer](#) | [Privacy](#) | [Copyright Notice](#)

© 2002 Medical College of Wisconsin